

3D Ultrasonographic Evaluation of the Fetal Heart with Ebstein's Anomaly: Original Image

Ebstein Anomalisi Olan Fetal Kalbin 3 Boyutlu Ultrasonografiyle Değerlendirilmesi

Selim BÜYÜKKURT, MD,^a
Nazan ÖZBARLAS, MD,^b
S. Cansun DEMİR, MD,^a
Fatma TUNCAY ÖZGÜNEN, MD,^a
Cüneyt EVRÜKE, MD,^a
Yusuf Taner KAFADAR, MD^a

Departments of

^aGynecology and Obstetrics,

^bPediatric Cardiology,

Çukurova University Faculty of Medicine,
Adana

Geliş Tarihi/Received: 21.09.2010

Kabul Tarihi/Accepted: 22.11.2010

Yazışma Adresi/Correspondence:

Selim BÜYÜKKURT, MD

Çukurova University Faculty of Medicine,
Department of Gynecology and Obstetrics,
Adana,

TÜRKİYE/TURKEY

selimbuyukkurt@hotmail.com

ABSTRACT Ebstein's anomaly is an infrequent form of congenital heart disease. The diagnosis can be made on 2D sonography by detecting the apical displacement of the septal and posterior leaflets of tricuspid. Valvular insufficiency and cardiomegaly are also commonly seen. A 3D examination of fetal Ebstein's anomaly is described at 21st week of gestation, showing the volumetric four chamber view of heart. 3D evaluation of fetal heart permits to better understand the anatomy as well as to store the volume in order to perform offline analysis.

Key Words: Ebstein anomaly; echocardiography, three-dimensional; prenatal diagnosis

ÖZET Ebstein anomalisi, konjenital kalp hastalıklarının nadir bir türüdür. İki boyutlu sonografide triküspidin septal ve arka kapaklarının apekse doğru yerleştiğinin saptanmasıyla tanısı konulabilir. Kapak yetmezliği ve kardiyomegali de sıklıkla görülür. Yirmi birinci gebelik haftasında kalbin hacimsel dört odacık görüntüsünün gösterildiği 3 boyutlu inceleme tarif edilmektedir. Fetal kalbin 3 boyutlu incelemesi anatominin daha iyi anlaşılmasını sağladığı gibi hacimsel bilginin saklanarak hasta kalktıktan sonra da incelemeye izin vermektedir.

Anahtar Kelimeler: Ebstein anomalisi; ekokardiyografi, üç boyutlu; prenatal tanı

Türkiye Klinikleri J Gynecol Obst 2011;21(1):39-41

Ebstein's anomaly is a rare form of congenital heart diseases (CHD) which accounts 0.3-0.6% of all cardiac defects.¹ The anomaly is characterised by the apical displacement of septal and posterior leaflets of tricuspid valve. The portion between the original atrioventricular valves and displaced cusps of the right ventricle is unified with right atrium which is called "atrialization of right ventricle". Tricuspid valve insufficiency, cardiomegaly and left axis due to right atrial dilatation are the classical findings.² Usually, the diagnosis could be made using the apical four chamber view on 2D sonography.

We examined a woman at age of 29 year old who was first admitted at the 21st week of gestation. She was the mother of a healthy girl at age of two years old and did not carry any risk factor for CHD. The examination was made using the convex volumetric transducer (RAB 4-8 MHz) on a VOLUSON 730 Pro (GE Medical Systems, Kretztechnik, Zipf, Austria) system.

Cardiomegaly was the initial finding causing the pulmonary hypoplasia. Apical four chamber view demonstrated the intraventricular attachment of posterior and septal leaflets of tricuspid. Color Doppler examination showed marked reflux during the ventricular systole (Figure 1). Volumes were acquired starting from the level of four chamber view of the heart where the sternum of the fetus is in closest position to the transducer. Volume acquisition and angle were set to low quality and 25° for scanning the entire volume of heart in one cardiac cycle. Multiplanar image was obtained and processed to get four chamber view on 3D (Figure 2). The family opted to terminate the pregnancy but denied to perform an autopsy.

Prognosis of fetal Ebstein's anomaly is usually lethal and a report of Polish cohort found that 90% of babies died mostly due to pulmonary hypoplasia.³ Berg et al. also reported a poor prognosis with Ebstein's anomaly. The authors pointed the importance of the difference of right atrial area index between the few survivors and the non survivors. While this difference did not found to be significant due to small number of cases, the alteration from the fetal to neonatal circulation interfere to determine the postnatal outcomes.⁴ In another report, Berg et al. had evaluated the hemodynamical changes in fetuses with obstructed right heart. They found that altered indices on the ductus venosus signify cardiac compromise in babies with Ebstein's anomaly in contrary to the other obstructive lesions of the right heart.⁵ However, these results require to further studies, due to lack of sufficient number of cases.

While the diagnosis of Ebstein's anomaly is usually possible using the 2D sonography, we believe that adding the 3D cardiac evaluation may permit to better demonstrate the CHD. The ability of storing the entire volume of heart is an other advantage of 3D allowing reevaluation.

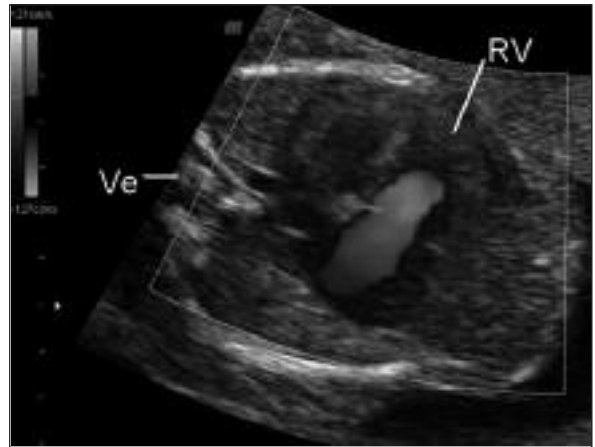


FIGURE 1: Apical four chamber view of the heart in color Doppler examination showing marked valvular insufficiency. Note the left axis of the heart due to right ventricular (RV) dilatation.

Ve: vertebra.

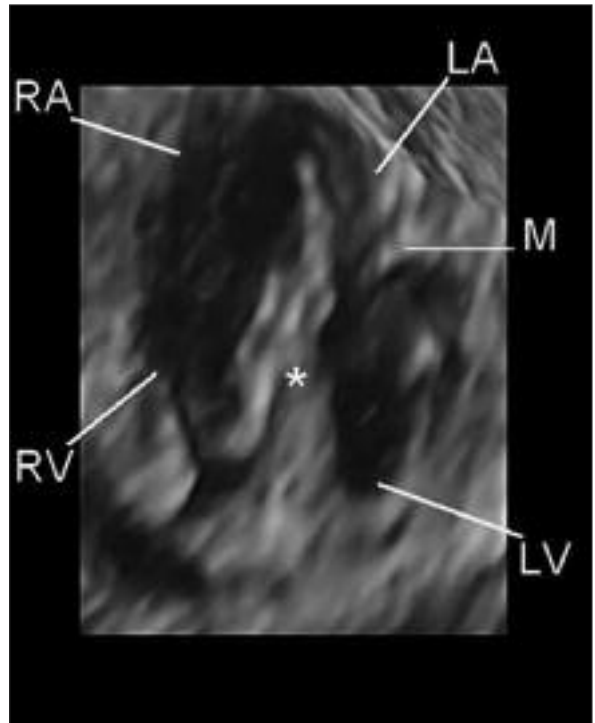


FIGURE 2: 3D four chamber view of the heart in Ebstein's anomaly. Asterisk located on the interventricular septum shows the low set insertion of the septal leaflet of tricuspid.

RA: right atrium, RV: right ventricle, LA: left atrium, LV: left ventricle, M: mitral valve.

REFERENCES

1. Starnes VA, Pitlick PT, Bernstein D, Griffin ML, Choy M, Shumway NE. Ebstein's anomaly appearing in the neonate. A new surgical approach. *J Thorac Cardiovasc Surg* 1991;101(6): 1082-7.
2. Melendres G, Ormsby EL, McGahan JP, Mon-Grady AJ, Towner D, Taylor D. Prenatal diagnosis of Ebstein anomaly: a potential pitfall. *J Ultrasound Med* 2004;23(4):551-5.
3. Respondek-Liberska M, Janiak K, Wilczynski J, Sysa A, Moll JA, Moll J, et al. Natural history of fetal Ebstein's anomaly in a referral center in the second half of pregnancy. *Ultrasound Obstet Gynecol* 2004;24(3):223-4.
4. Berg C, Geipel A, Kohl T, Bender F, Breuer J, Herberg U, et al. Ebstein's malformation detected in fetal life - associated findings, intra-uterine course and outcome. *Ultrasound Obstet Gynecol* 2006;28(4):441.
5. Berg C, Kremer C, Geipel A, Kohl T, Germer U, Gembruch U. Ductus venosus blood flow alterations in fetuses with obstructive lesions of the right heart. *Ultrasound Obstet Gynecol* 2006;28(2):137-42.